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Subjective visual vertical in Pisa syndrome

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ABSTRACT

Background: Parkinson's Disease (PD) alters perception and somatosensory information integration, including visual dependency and judgment of body position in space. PD may be associated with Pisa syndrome (PS), a lateral deviation of the longitudinal body axis (LBA) of unknown origin. We tested whether this inclination is associated with an altered perception of the subjective visual vertical (SVV) and if these alterations are secondary effects of the LBA deviation or of a primary perceptual dysfunction. Furthermore, we investigated the contribution of different sensory modalities and dopaminergic medication.

Methods: Seventeen PD patients (8 with PS, 9 without PS) and 18 healthy controls were tested. The SVV was assessed in a seated, in a lateral horizontal and – in PS patients – in a seated manually rectified position. Frame and moving-stimulus-patterns were used to test visual dependency. In PD and PS patients all trials were conducted in dopaminergic "on" and "off".

Results: When seated, SVV values on PD in "on" and PS in "on" and "off" differed significantly from controls. This difference remained in PS patients after manual rectification in "off". The SVV in a lateral horizontal position was not significantly different between the three groups. When inclined, visual dependency was higher in PD "off" than in controls.

Discussion: Both PS and PD patients showed SVV deviations compared to healthy controls. These cannot be explained by their intrinsic lateral deviation in PS patients. They must be secondary to either a primary perceptual dysfunction or alterations of internal models of verticality due to re-weighting of perceptual afferences.

1. Introduction

Parkinson's disease (PD) is a neurodegenerative disease resulting mainly from loss of midbrain dopaminergic neurons. Originally characterized as a motor disorder, it is also known to affect perception and somatosensory information integration, such as proprioceptive feedback information of static position and movement perception [1]. Deficits in orientation and postural stabilization secondary to disturbances of somatosensory processing have been shown in PD when angular acceleration information coming from the vestibular system is isolated [2]. Patients with PD have been shown to rely more on visual information when performing tasks involving arm movements [3] and postural orientation and stabilization [2]. It has been proposed that the enhanced visual dependency in PD for orientation and postural stabilization may result from an attempt to compensate for impaired somatosensory processing [2].

The perception of orientation in space involves multiple sensory modalities, corresponding to visual, somatosensory and vestibular systems. The perception of earth-verticality in the roll plane (i.e. around the naso-occipital axis) has been studied extensively with psychophysical tasks such as the subjective visual vertical (SVV). This task is affected by gravitational forces which are perceived by the otolithic vestibular system, visual references to verticality cues, and the perceived longitudinal body axis (LBA). A widely accepted model proposes that the SVV results from the vectorial summation of the weighted influences of these three factors [4] (Fig. 1A).

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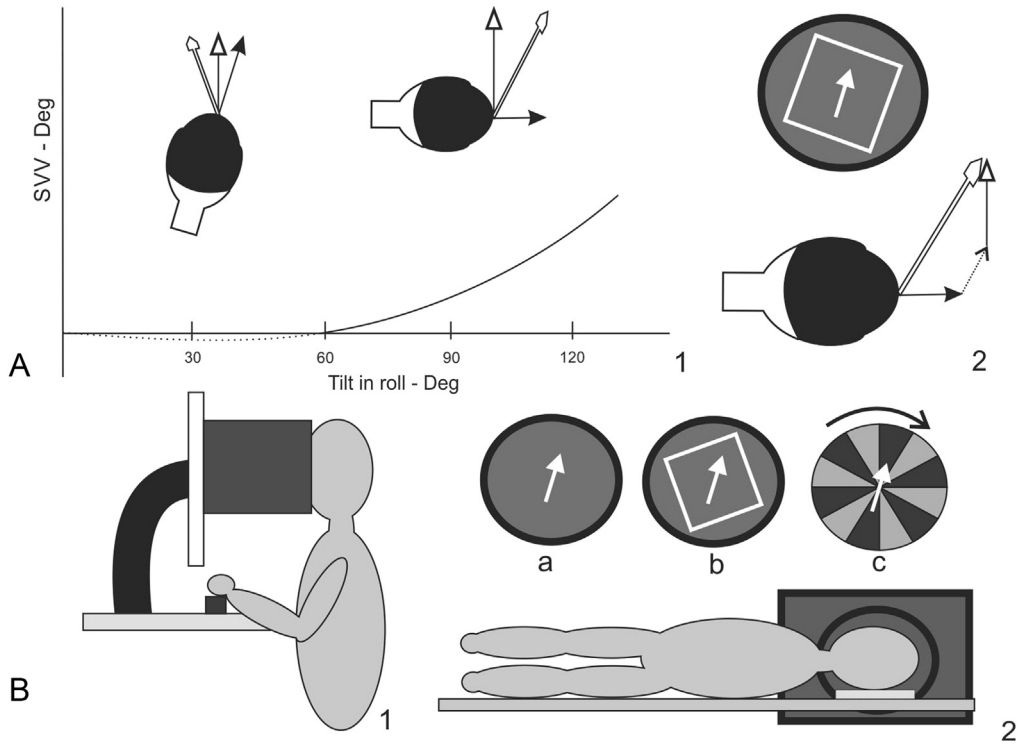

Fig. 1. (A) (1) Dependency of the SVV deviation from the true gravitational vertical on the roll tilt of the longitudinal body axis (LBA). Dotted line represents the E-effect and black line the A-effect. The black arrow represents the LBA, the white arrowhead the otholitic afference, and the empty arrow the resultant SVV. 2-SVV modeled as vectorial summation of LBA, otholitic afferences, and visual verticality cues (dotted arrow). (B) Diagram of the device used for SVV determination. (1) Seated position; and (2) right lateral decubitus position. a, b, c correspond to black, frame and optokinetic backgound conditions.

In the absence of vestibular or CNS disease, subjects are very accurate in this task, usually setting the line within less than two degrees of the gravitational vertical. Tilts of the SVV can be observed in vestibular disease or CNS disorders involving vestibular pathways [5]. However, large SVV tilts can be induced even in normal subjects: when they are lying >60° to one side they tend to set the SVV tilted the LBA direction inclination by some 10–30°. This is called the "Aubert" or "A" effect [6], which is thought to be a compromise between a tendency to set the line to the true gravitational vertical and a trend to set the line parallel to the LBA [7]. Another - less frequent phenomenon - may be encountered, when subjects are inclined less than 60° in the roll plane. They tend to overcompensate and set the line tilted to the opposite direction of body tilt (E-effect) [8] (Fig. 1A).

Patients with PD differ significantly from controls as to how vestibular, visual and propioceptive information are weighted for verticality perception. PD patients are more dependent on visual cues in SVV trials and other spatial tasks related to perceived orientation [9]. The significance of vestibular input in PD is controversial. Some studies claim that PD patients are more reliant on vestibular information than on their perception of the LBA [10] and that vestibular dysfunction cannot explain the postural deficits of patients who are mildly or moderately affected by PD [11]. However, vestibular deficits as well as reduced or even absent vestibulo-collic reflexes have been described in PD patients [12].

Dopaminergic medication increases the weight of vestibular and visual information in verticality perception. This effect may be caused by altered somatic information processing as there is evidence that dopaminergic stimuli deteriorate proprioceptive information processing [10].

Pisa syndrome (PS) is an uncommon syndrome manifested by a significant lateral deviation of the body axis. Its presentation can be acute or insidious and it has been associated with the use of different CNS-active drugs and with neurodegenerative diseases such as Alzheimer disease, Multiple System Atrophy, and Parkinson's disease. The pathophysiology of PS and camptocormia (abnormal forward flexion of the spine) is still obscure. A cholinergic-dopaminergic imbalance [13], as well as mechanical causes related to axial myopathy [14] or dystonic flexion of the para-spinal muscles [15] have been proposed as possible causes.

Personal clinical observations such as the absence of a counter-rotating head posture to compensate for the thoracic inclination and resistance to attempts at obtaining an upright position by passive modification of the posture suggest that the abnormal body position in PS may be due to a perceptual alteration of verticality.

In the present study we attempt to elucidate whether the lateral inclination of patients with Pisa syndrome is associated with an altered perception of SVV. We also try to clarify whether this alteration is due to a manifestation of the A- or E-effect, given the inner roll-plane deviation of these patients, and/or a primary perceptual dysfunction. Additionally, we set out to determine the contribution of different sensory modalities (vision, graviceptive-somatoperception, otolithic input) and dopaminergic modulation on verticality perception in this population.

2. Patients and methods

2.1. Patients

Subjects were recruited from the out-patient clinic of the Movement Disorders Unit at the Institute of Neurosciences, Favaloro Foundation University Hospital in Buenos Aires, Argentina. 17 patients (9 male, 8 female) diagnosed with idiopathic PD according to the UK Brain Bank Criteria [16] were included. Eight suffered from Pisa

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